Retroperitoneal fibrosis

Inflammatory fibrotic process in the retroperitoneum causing compression of retroperitoneal structures, including ureters.

Incidence 1:200,000

Peak age 40-60

Males > females 2:1

Aetiology

- **Idiopathic (70%)** — ? immune response to ceroid, a polymer of oxidised lipids and protein from atherosclerotic plaques
- Drugs
  - Methysergide* — migraine treatment
  - LSD*
  - Bromocriptine*
  - Methyldopa* — *all ergot alkaloids
  - Beta blockers
  - Phenacetin
  - Amphetamine
- Malignancy
  - Lymphoma
  - Sarcoma
  - Carcinoma of breast, colon, stomach and prostate
- Infection
  - TB
  - Schistosomiasis
  - Chronic UTI
  - Syphyllis
  - Gonorrhoea
  - *Actinomyces*
- Radiation
- Inflammatory
  - IBD
  - Sarcoidosis
  - Endometriosis
  - Collagen diseases
  - Inflammatory AAA
  - Multifocal fibrosclerosis — rare condition characterised by RPF mediastinal and mesenteric fibrosis sclerosing cholangitis, Reidel's thyoiditis and orbital pseudotumour

Presentation

Relatively non-specific often with delayed diagnosis

- Vague back pain
- Lethargy
- Anorexia
- Weight loss
- Symptoms attributable to disease process
  - Ureters — oliguria, weight gain, SOB
  - IVC — DVT
  - Renal vein — hypertension, haematuria

Pathology

Smooth, flat tan-coloured mass of tissue overlying ureters

Histology	Early phase	collagen and inflammatory cells
	Late phase	collagen
	Malignancy RPF	collagen and occasional islands of tumour cells

Diagnosis

Raised ESR, CRP, anaemia and leucocytosis common but non-specific
No current diagnostic serological test
IVU shows varing degree of obstruction with medial deviation of ureters (NB. medial deviation seen in ~20% of normal individuals)
CT or MRI with or without contrast are the investigations of choice
Except in a few obvious cases of lymphoma, cross-sectional imaging not sufficient to exclude malignancy: Campbell's states that:
'Representative **biopsies of the mass need to be obtained** ..to rule out malignancy and allow one to proceed with treatment for RPF' However some believe that biopsy not required if:

- Classic features of RPF on CT/MRI
- No lymphadenopathy
- No history of prior malignancy
- ? a role for PET scanning in this situation to exclude requirement for biopsy - both lymphoma and sarcoma positive on FDG-PET

Management

Initial managent comprises decompression of urinary tracts or Mx of DVT
Primary high dose steroid therapy

- Effective in ~80%
- ?better in those with high ESR, leucocytosis or inflammatory cells on biopsy
- Duration of treatment unknown; > 6 months recommended, but because of relapse rate (?value) some advocate longer term Rx
- Rx schedule (Kardar 2002) in Campbell's:

> (ESR). The steroids were started with an initial oral intake of 60 mg. prednisolone on alternate days for 2 months. This dose was tapered during the next 2 months (40 mg. for 2 weeks, 20 mg. for 2 weeks and 10 mg. for 2 weeks) to a maintenance dose of 5 mg. daily. Total duration of steroid use was 2 years.

Steroid-sparing immunosuppressants including azathioprine, cyscosporine, mycophenylate mofetil and tamoxifen all reported to have efficacy. Possible role for non-steroid immunosuppressants in preventing relapse after short-course of high-dose steroids (Swartz RD 2009)
Surgical management for non-responders:

- Open ureterolysis
  - Midline incision
  - Medial mobilisation of left and right colon
  - Initial biopsy +/- frozen section
  - Right-angle from normal ureter to abnormal

- Placement of ureters in peritoneum or omental wrap
- If inadvertent uretotomy leave stents longer post-op

Laparoscopic ureterolyis

- First report Clayman and Kavoussi 1992
- Data on 13 patients reported in 2002
- All pre-stented and placed in perioneum by tacking white line back to original site *underneath* ureter
- Conversion in 15% for bleeding/failure to progress; no intrabdominal complications; 92% unobstructed at 30 months